Radiological Manifestations of Fallot's Tetralogy

THESIS

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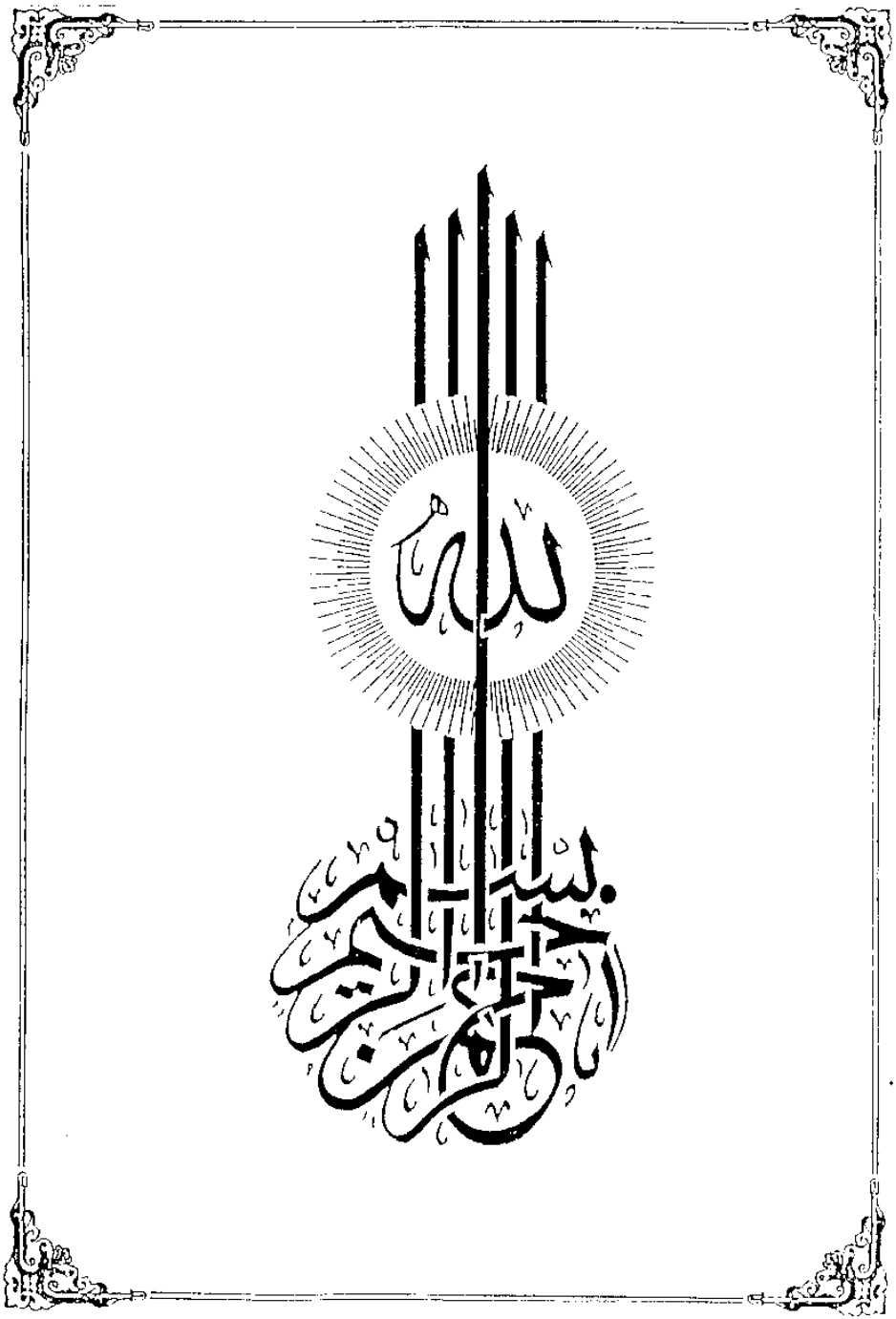

ETIENNE-LOUIS-ARTHUR FALLOT
(1850 - 1911)

CONTENTS

INTRODUTION AND AIM OF WORK

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The first description of the tetralogy of Fallot was by Niels Stensen (1638-1686) in 1671. It was not until 1888 that Etienne-Louis-Arthur Fallot (1850-1911) presented the most comprehensive account of "maladie bleue" that was to bear his name even though it had clearly been described before by several authors (Willius & Dry, 1948).

Fallot's important work appeared under the title, "*Contribution à l'anatomie pathologique de la maladie bleue (cyanose cardiaque)* " in 1888. The ensuing quotation, in translation, partly records his conclusions.

> This malformation consists of a true anatomopathologic type represented by the following : (1) Stenosis of the pulmonary artery ; (2) Interventricular communication ; (3) Deviation of the origin of the aorta to the right ; (4) Hypertrophy, almost always concentric , of the right ventricle. Failure of obliteration of the foramen ovale may occasionally be added in a wholly accessory manner.

Tetralogy of Fallot is a relatively common congenital heart disease which constitutes approximately 5 to 10% of infants and children with congenital defects. It is the most common single lesion in patients with cyanotic disease who live beyond infancy; it has been estimated that if the diagnosis of tetralogy of Fallot were made in every cyanotic child with heart disease who was more than two years old it would be correct at least 75% of the time (Nadas & Fyler, 1972).

The surgical intervention in this anomaly did not come until 1944 when Dr. Helen Taussig together with Alfred Blalock devised the subclavian artery-to-pulmonary artery anastomosis (Blalock & Taussig, 1945).

The first intracardiac repair on a child with tetralogy was performed by C. Walton Lillehei in 1954, using cross circulation from a donor (Lillehei *et al.* , 1955).

The converging forces of surgical innovation and technological development are stimulating rapid progress in the diagnosis and treatment of patients with congenital heart disease. The trend in cardiac surgery toward complete repair of tetralogy of Fallot in infancy (Castaneda *et al.* , 1977) has necessitated accurate and complete diagnosis in the first months of life. Concomitant advances in electronics and computerization are facilitating new (and it is hoped, more accurate) means of diagnosis to meet this challenge.

This work is intended to present the current status of the different imaging modalities used in the diagnosis of Fallot's tetralogy.

Improvement in diagnosis received a big boost in the past decade from two areas, cineangiography and two-dimensional echocardiography, and those innovations by the introduction of axillary angled views by Bargeron and his associates in 1977. Also the creation of new catheters for enlarging or occluding cardiovascular structures at the time of cardiac catheterization has widened the horizons for treatment of many lesions. A look at the future and into the new modalities of digital angiocardiography, computed tomography, nuclear cardiology, and magnetic resonance imaging will be provided.

ANATOMICAL CONSIDERATIONS

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1. NORMAL ANATOMY OF CARDIAC CHAMBERS

The long axis of the heart, from base to apex, runs downwards, anteriorly and to the left. The right atrium and right ventricle occupy the anterior portion of the heart and the left atrium and ventricle lie posteriorly.

Because of the anterior position of the right heart chambers, the aorta and its valve have a central position in the heart, being wedged between the atrioventricular valves and posterior to the infundibulum of the right ventricle (Macartney, 1983).

THE RIGHT ATRIUM

In the normal heart this chamber receives the systemic venous return. The superior vena cava, inferior vena cava, and coronary sinus enter the smooth walled sinus venarum, which is separated from the trabeculated right atrial appendage by the prominent crista terminalis. On its septal surface the right atrium exhibits the limbus of the fossa ovalis .

THE LEFT ATRIUM

This chamber receives the pulmonary venous return through four pulmonary veins. These veins enter a smooth walled segment which is not separated from the trabeculated atrial appendage by a crista. On its septal aspect, the left atrium presents the flap valve of the septum primum .

THE RIGHT VENTRICLE

The right ventricle is triangular in shape and forms a crescentic, shallow structure wrapped over the ventricular septum. It can be divided, as shown in **figure 1**, into an "inlet" portion, such surrounds and supports the tricuspid valve, a "trabecular" portion, and an "infundibulum" or outlet portion, which is a muscular funnel that gives attachement to the pulmonary valve (Anderson & Becker, 1983). According to Van Mierop (1974), the right ventricular cavity can also be divided into a posteroinferior inflow portion, and an anterosuperior outflow portion.

The term "crista supraventricularis" describes the supraventricular muscle mass that separates the pulmonary and tricuspid valves **(Figs 2 & 3a)**. It achieves two anatomic functions (i) it separates tricuspid and pulmonary valves and (ii) separates the outflow tracts of the two ventricles (Macartney, 1983).

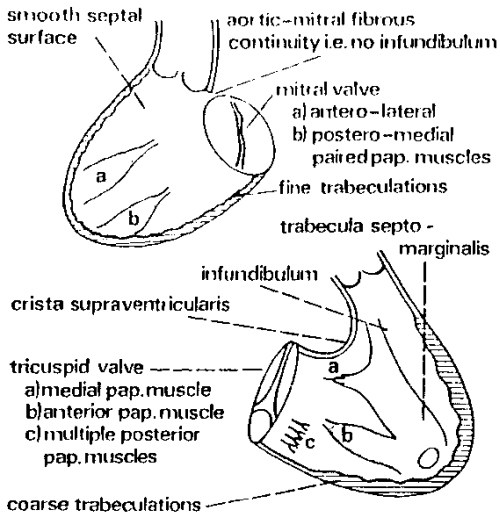

Fig. 1. Diagram illustrating the morphological differences between the normal right ventricle (**bottom**) and left ventricle (**top**). pap. = papillary. [From Macartney, 1983.]

Fig. 2. Normal right ventriculogram, right anterior oblique view. The normal crista supraventricularis is an unbroken line from its junction with the tricuspid valve ring (**lower arrow**) to its junction with the pulmonary valve ring (**upper arrow**). [From Partridge & Fiddler, 1981.]

The crista supraventricularis is formed of two components (Anderson, Becker & Van Mierop, 1977), the larger part is made of the inner curvature of the heart wall. This is the "ventriculo-infundibular fold" , and incisions through this part open outside the heart **(Fig.3b)**. Only the most septal part of the crista is between the pulmonary and aortic outflow tracts. This is the "infundibular septum" (the parietal band is an alternative term at present in widespread usage). Its septal part is inserted between the limbs of the trabecula septomarginalis, while its parietal part imperceptibly merges with the ventriculo-infundibular fold **(Fig.3b)**.

The two components of the crista can be divorced in hearts with conotruncal anomalies (e.g., tetralogy of Fallot) ; and instead of one muscle band, the crista, there are two : one the ventriculo-infundibular fold; the other the infundibular septum. For this reason, Anderson *et al.* (1977) submit that the term crista supraventricularis should be used only in the description of hearts with normal right ventricular outflow tracts.

The "trabecula septomarginalis" is a prominent Y-shaped muscle band on the right ventricular septal surface. Its anterior limb runs upwards toward the pulmonary valve while the posterior limb extends into the inlet portion. The body of the trabecula septomarginalis extends apically to become continuous with the "moderator band" , a prominent trabeculation running from septum to free wall **(Fig.1)**.

Some authorities categorise the trabecula septomarginalis as the "septal band " of the crista, but the trabecula is purely a septal structure and in no way a supraventricular crest (Anderson *et al.* , 1974). Actually, the ventriculo-infundibular fold, infundibular septum and trabecula septomarginalis have an entirely different embryologic origin; they just happen to become lined up in normal development but do not necessarily do so in anomalous hearts (Van Mierop, 1974).

The three tricuspid leaflets are positioned septally, antero-superiorly and inferiorly **(Fig.1)**. A large "anterior papillary muscle" supports the antero-superior leaflet and fuses with the moderator band at the free wall. Multiple smaller "posterior papillary muscles" take origin from the apical part of the trabecula septomarginalis and support the inferior and septal leaflets. There is a group of small septal papillary muscles; the uppermost one, called the "medial (conal) papillary muscle" (muscle of Lancisi), is attached to the posterior limb of the trabecula septomarginalis.

THE LEFT VENTRICLE

The main features which differentiate the left ventricle from the right ventricle **(Fig.1)** are firstly that its inlet and outlet valves are in "fibrous continuity" ; secondly, that the trabeculations of the left ventricle are fine; and finally, that the septal surface is smooth, lacking any structure comparable with the trabecula septomarginalis of the right ventricle (Macartney, 1983).

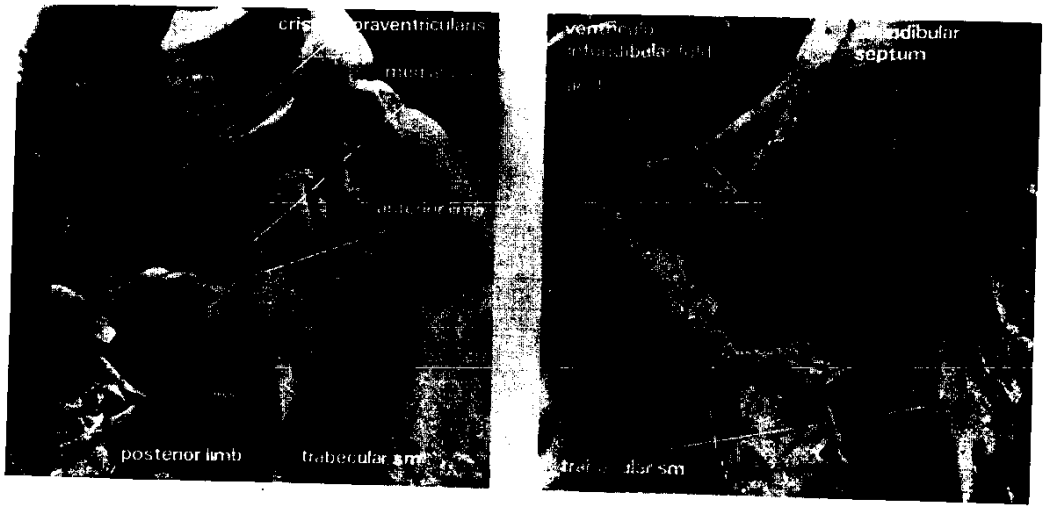

Fig. 3. Photographs of normal heart taken as if looking from the right ventricular apex directly into the pulmonary outflow tract. (**A**) The normal crista supraventricularis separates the tricuspid and pulmonary valves. (**B**) Dissection of crista shows that the greater part, the ventriculo-infundibular fold, is merging with the infundibular septum which is inserted between the limbs of the trabecula septomarginalis. [From Anderson & Becker, 1983.]

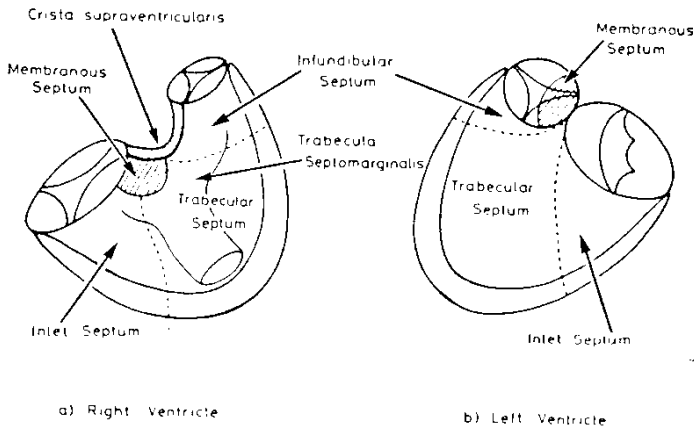

Fig. 4. Diagram illustrating the different muscular components of the ventricular septum and their conjunction with the membranous septum. [From Soto *et al.* , 1980.]

The "inlet" portion extends from the mitral valve annulus to the attachment of the papillary muscles and contains the mitral valve. This valve has two leaflets, an anterior or septal leaflet and a posterior or mural leaflet, separated by antero-lateral and postero-medial commissures, and is supported by two papillary muscles, again in postero-medial and antero-lateral positions.

The "trabecular zone" of the ventricle extends from the attachments of the papillary muscles to the ventricular apex. The "outflow" tract of the ventricle extends from the trabecular zone to the aortic valve. Posteriorly, the anterior mitral leaflet separates the inlet and outlet portions.

The aortic valve forms the distal margin of the left ventricular outflow tract and has three leaflets : the right coronary leaflet in front, the left coronary leaflet on the left and non-coronary leaflet posteriorly and to the right (Anderson & Becker, 1983).

THE INTERVENTRICULAR SEPTUM

This is formed by two structures : the "membranous" and the "muscular" components (Soto *et al.* , 1980). The membranous septum is a very small fibrous structure, a component of the central fibrous body, abutting with the aortic, mitral, and tricuspid valve annuli. It has two parts divided by the implantation of the central part of the tricuspid-septal leaflet. They are : (1) the "atrioventricular" component, located above the septal tricuspid leaflet implantation, and (2) the "interventricular" component, located below such implantation. The atrioventricular component of the membranous septum, which constitutes most of its area, separates a small part of the right atrium from the left ventricle.

The major part of the ventricular septum is muscular and has been divided into three segments : inlet, outlet, and trabecular **(Fig.4)**. The "inlet septum" is related with the attachment of the tricuspid-septal leaflet on the right side and the mitral-septal leaflet on the left side. The "outlet" or "infundibular septum" separates the outflow tracts of the ventricles and is located immediately beneath the arterial valves. The "trabecular septum" represents the more extensive part of the muscular septum and separates the trabecular components of both ventricles (Soto *et al.* , 1980).

The inlet septum is nearly at right angles to the outlet septum. It is more or less in the sagittal plane of the body, while the outlet septum is more frontally oriented (Anderson & Becker, 1983) (see Fig.64).

2. EMBRYOLOGY OF TETRALOGY OF FALLOT

FORMATION OF THE HEART LOOP

The heart is essentially a straight tube that lies within the pericardial cavity. From this tube will develop the embryonic ventricle and bulbus cordis and may , therefore, be called the "bulboventricular tube" **(Fig.5A)**.

As it grows the bulboventricular tube bends rightward and anteriorly, initially into the shape of the letter C , and later into a compound sigmoid structure - the "bulboventricular loop" . The deepening concavity on the left side of the bulboventricular loop is referred to as the "bulboventricular or conoventricular groove". This groove corresponds internally to a fold, " the inner heart curvature" or "bulboventricular or conoventricular flange" or "bulboatrioventricular ledge" **(Fig.5)** (Van Mierop, 1979).

At an ovulation age of about 25 days, the heart completely occupies the pericardial cavity, with the primitive left ventricle on the left side and the bulbus cordis on the right. The junction between the ventricle and the proximal bulbus cordis is called the "primary interventricular foramen" .

The proximal one-third of the bulbus cordis eventually will form most of the body of the right ventricle and may, therefore, be called "the primitive right ventricle ". From the middle one-third of the bulbus - the "conus cordis" - the outflow portions of both ventricles will be derived. The terminal one-third of the bulbus after partitioning, develops into the aortic and pulmonary roots and may therefore be called the "truncus arteriosus" (Van Mierop, 1979).

NORMAL CONAL INVERSION AND CONAL ABSORPTION

A vital feature of normal embryogenesis is the normal inversion of the conus (Goor, Dische & Lillehei, 1972). This process is dependent upon looping of the bulboventricular tube and is reflected by " the position of the conal septum " . This structure possesses septal and parietal insertions which correspond to the positions of the conal ridges. Thus, the septal insertion is sinistro(left)-anterior and the parietal insertion is dextro(right)-posterior **(Fig.6A)** (Becker, Connor & Anderson , 1975).

Normal conal inversion does not produce a situation in which the aorta is above the left ventricle. Instead, it produces a situation in which the aorta is posterior and slightly to the right to the pulmonary artery (dextro-posterior position) **(Fig.6A)** . At this stage the conoventricular flange separates the developing semilunar and atrioventricular valves.